

Fig. 1a Non-Transduced hMSCs in Immuno-competent
mouse muscle
Day 3
Dil + hMSCs

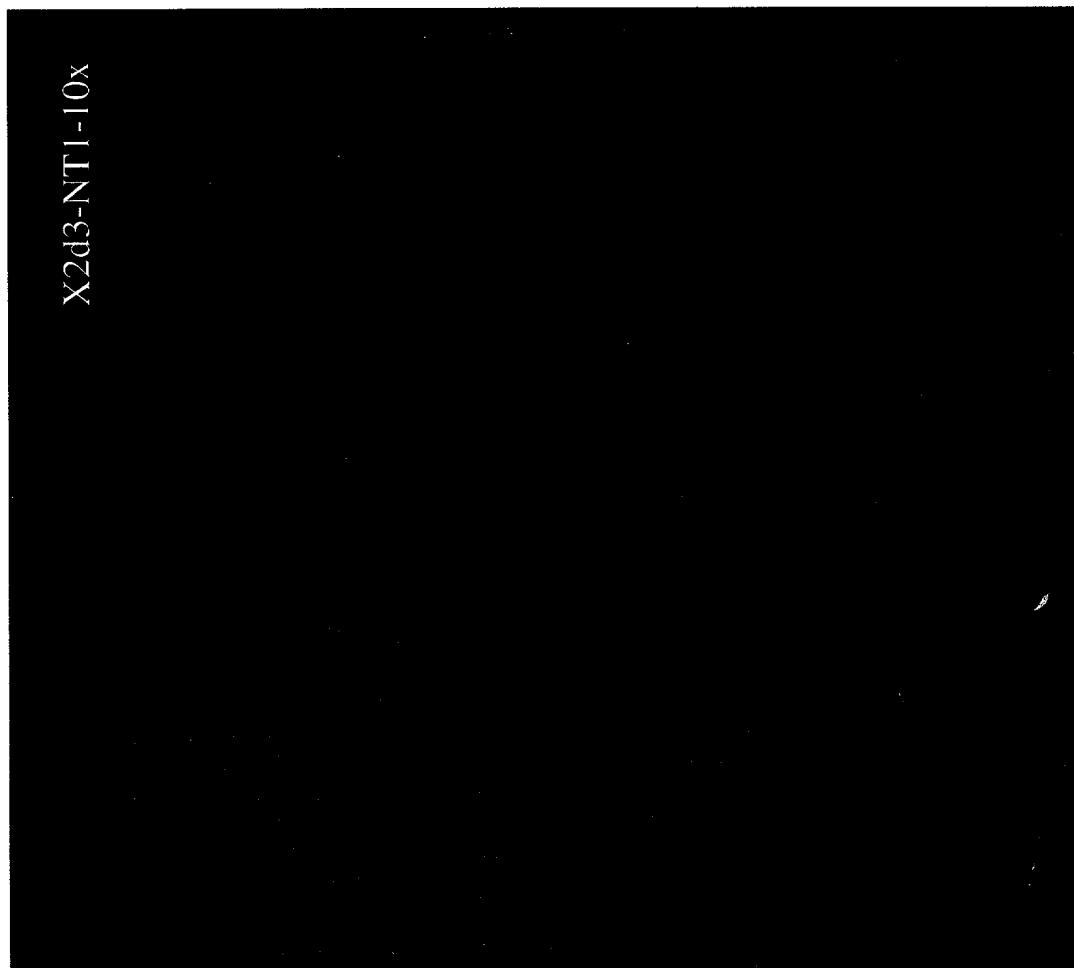
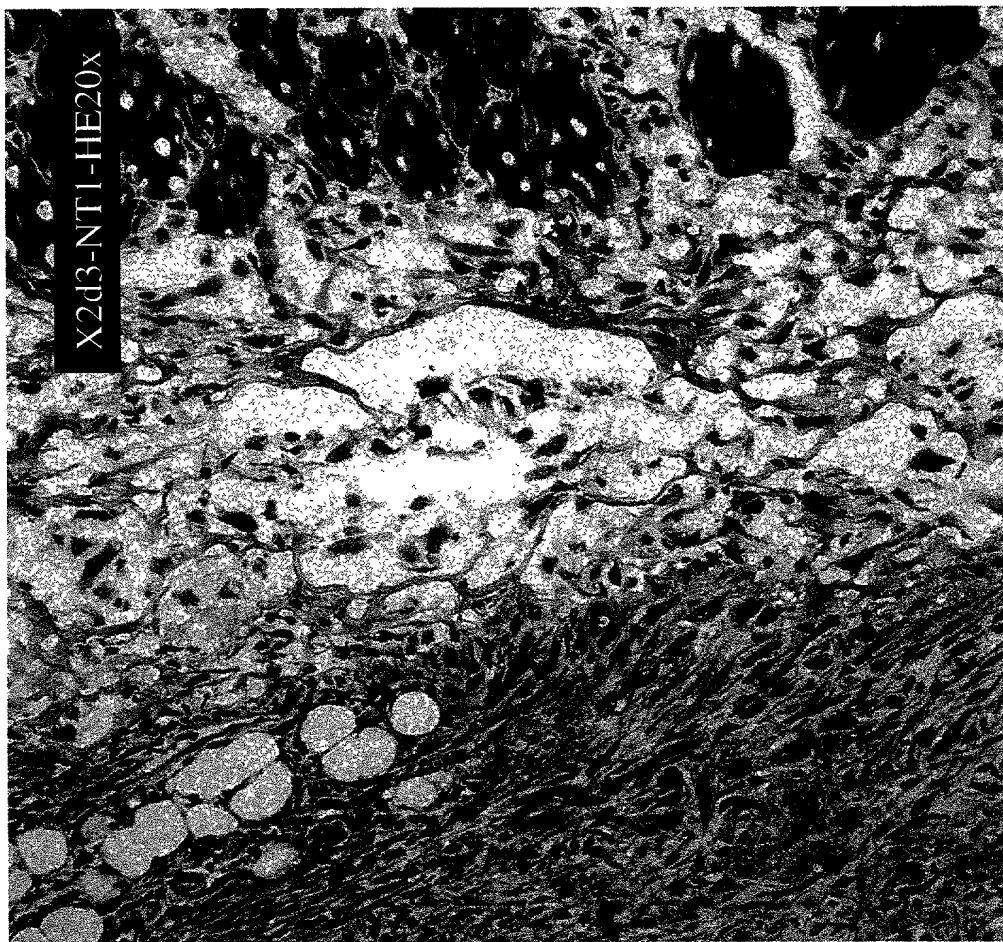
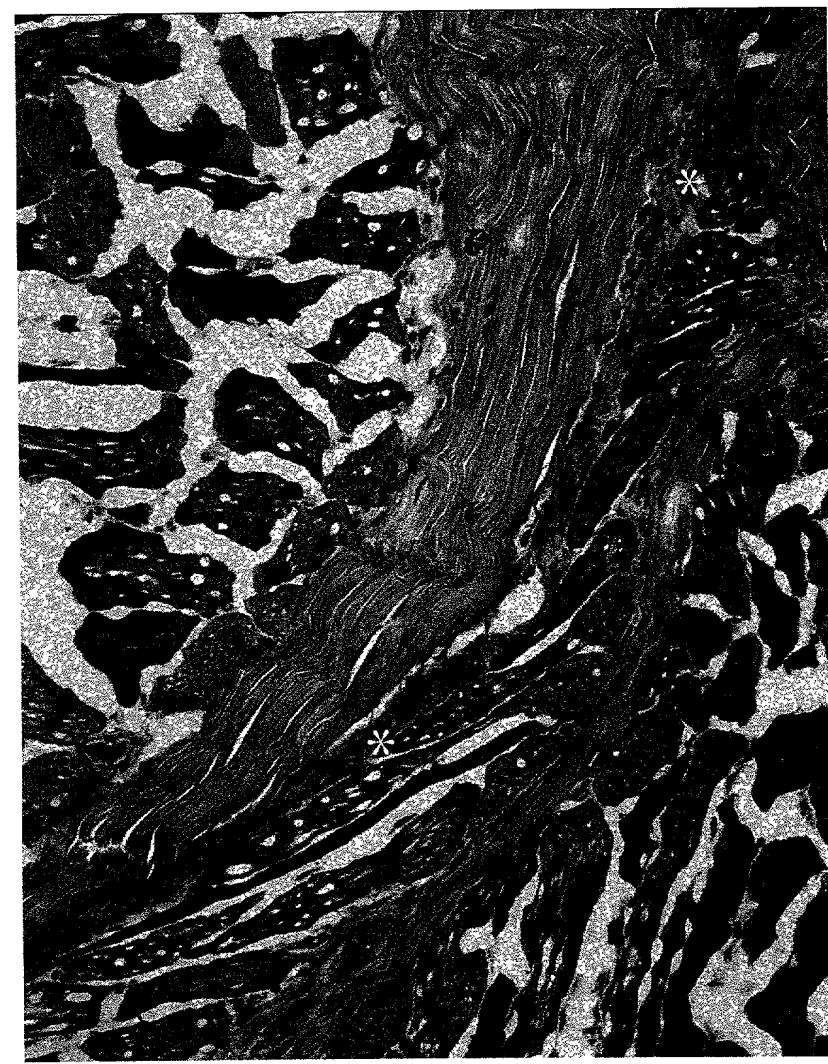


Fig.1b

Human MSCs in Mouse Muscle: Day-7

(H&E)



Dil labelled cells +
Sybr-green staining
of all nuclei

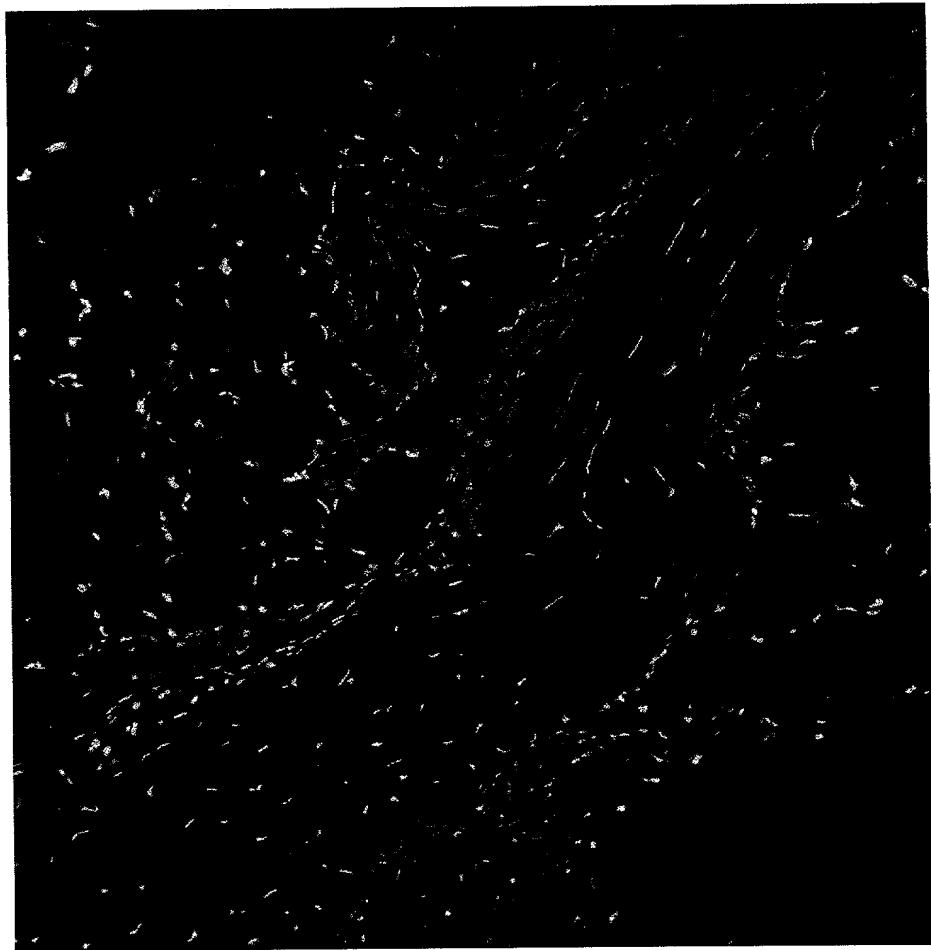


Fig.1c Human MSCs in Mouse Muscle: Day-7

(H&E, 20X)

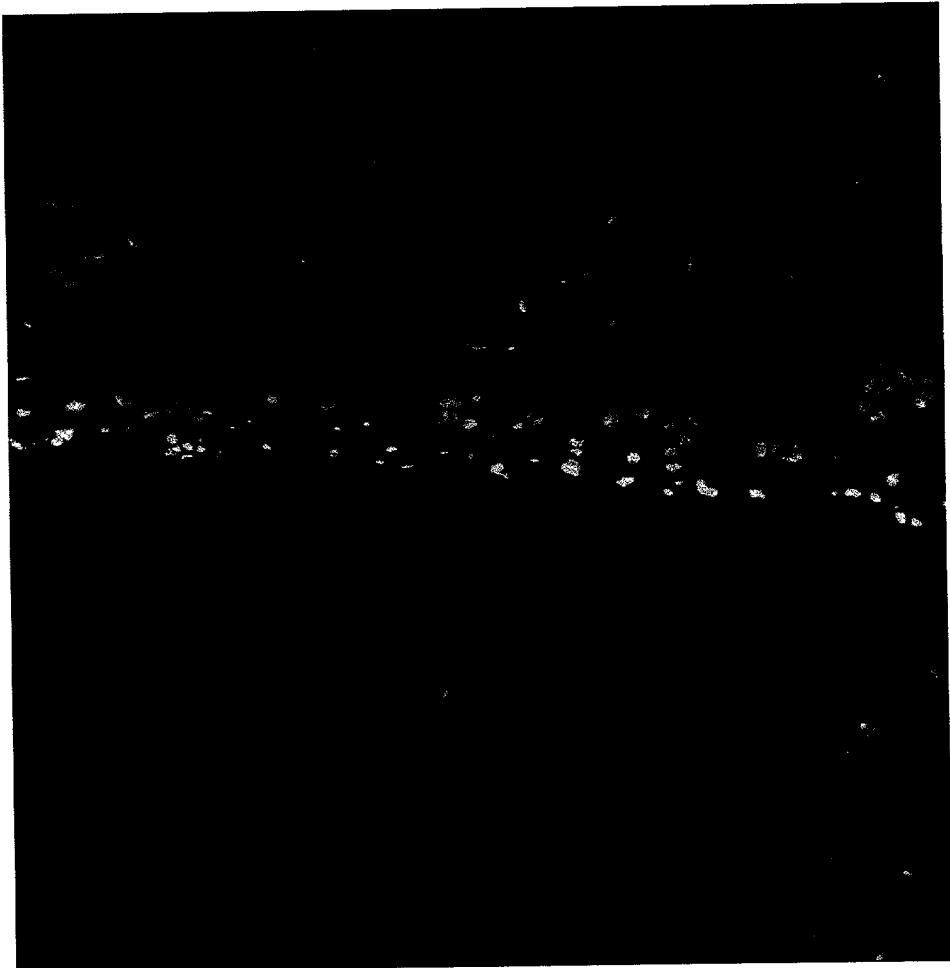
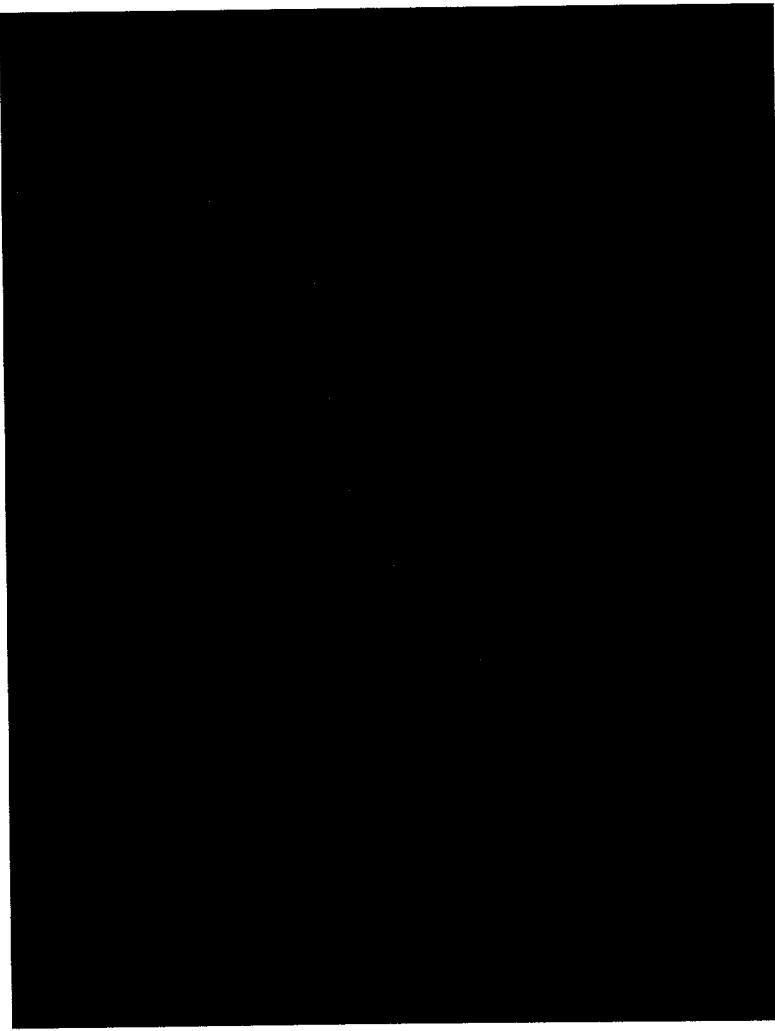


Fig. 1d

Human MSCs in Mouse Muscle: Day 14

Fluorescent Field, 10X



H&E 40X

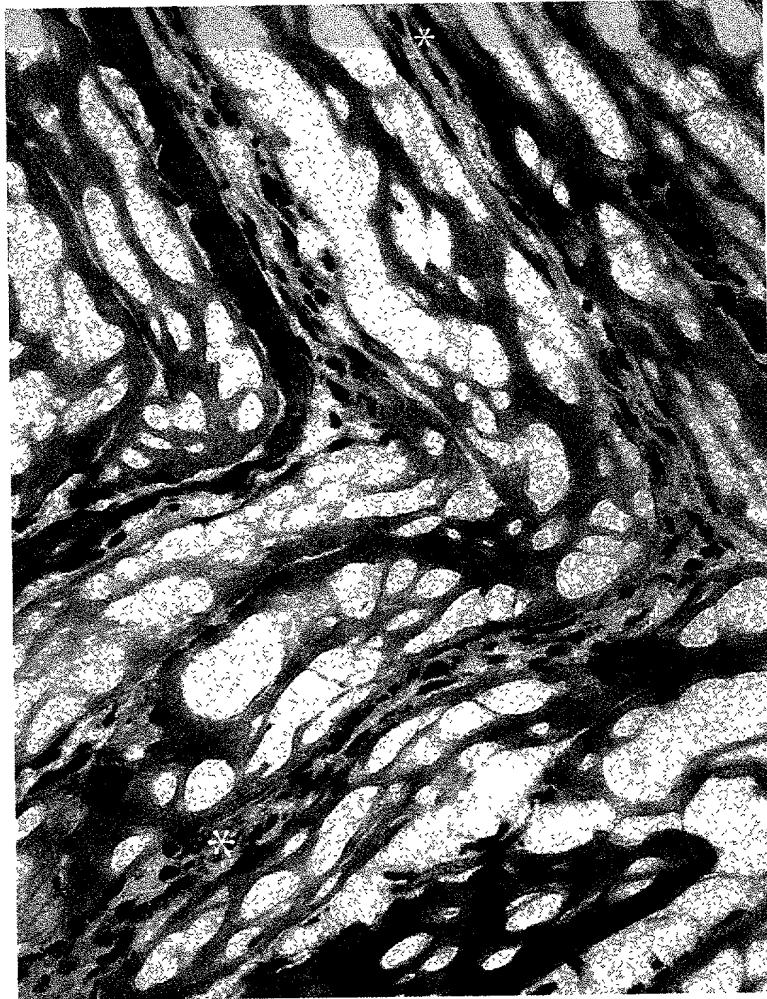


Fig. 1e Dil⁺ Positive α -GalA Transduced hMSCs in

Immuno-competent mouse muscle

d29

d43

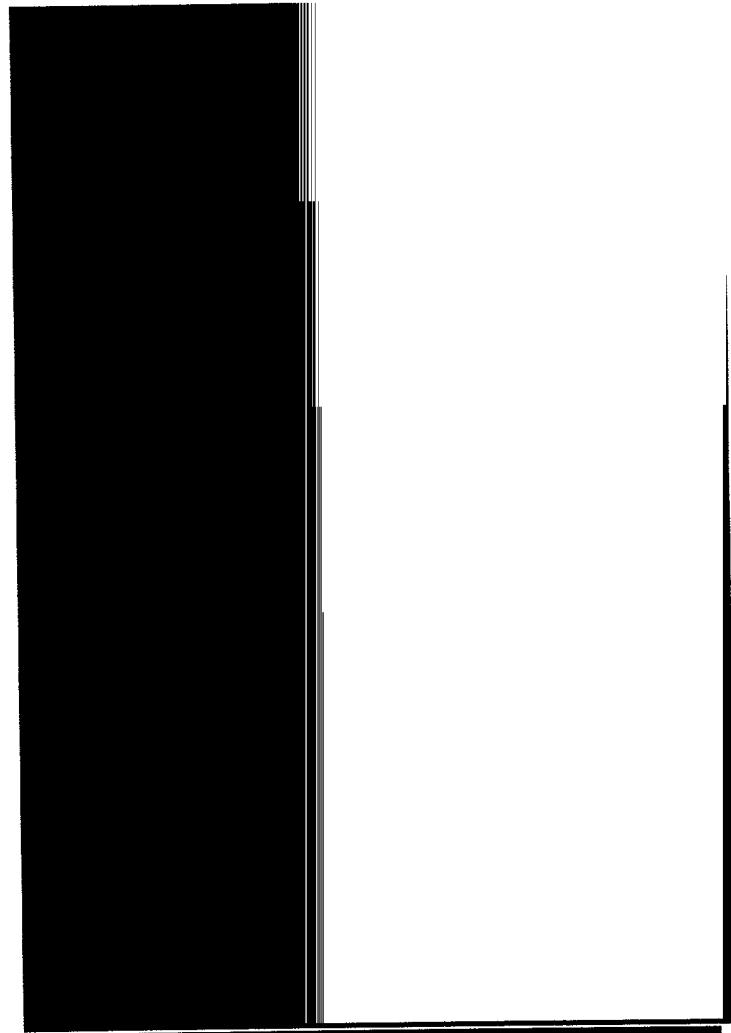
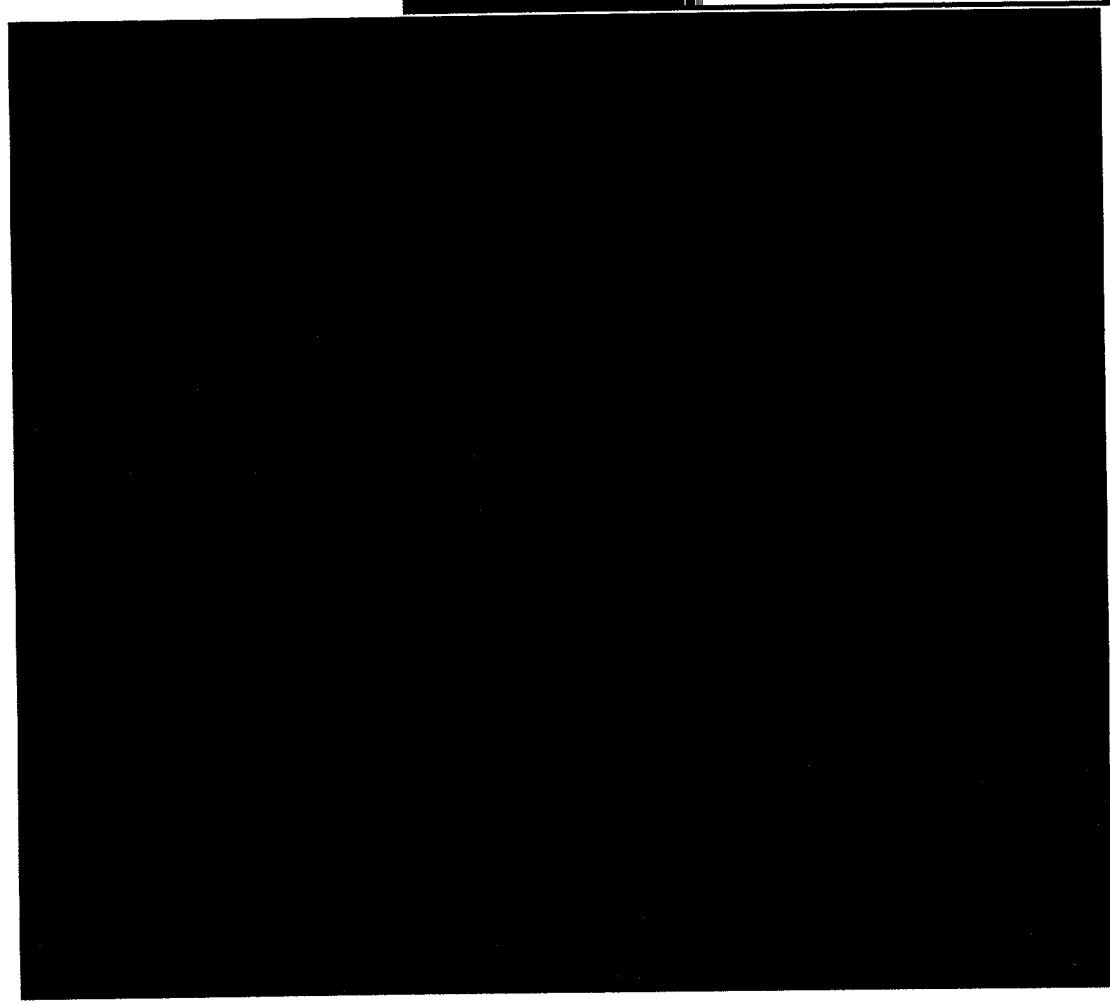


Fig. 1f

Human α GalA-Transduced MSCs in Mouse Muscle: Day-57

10X

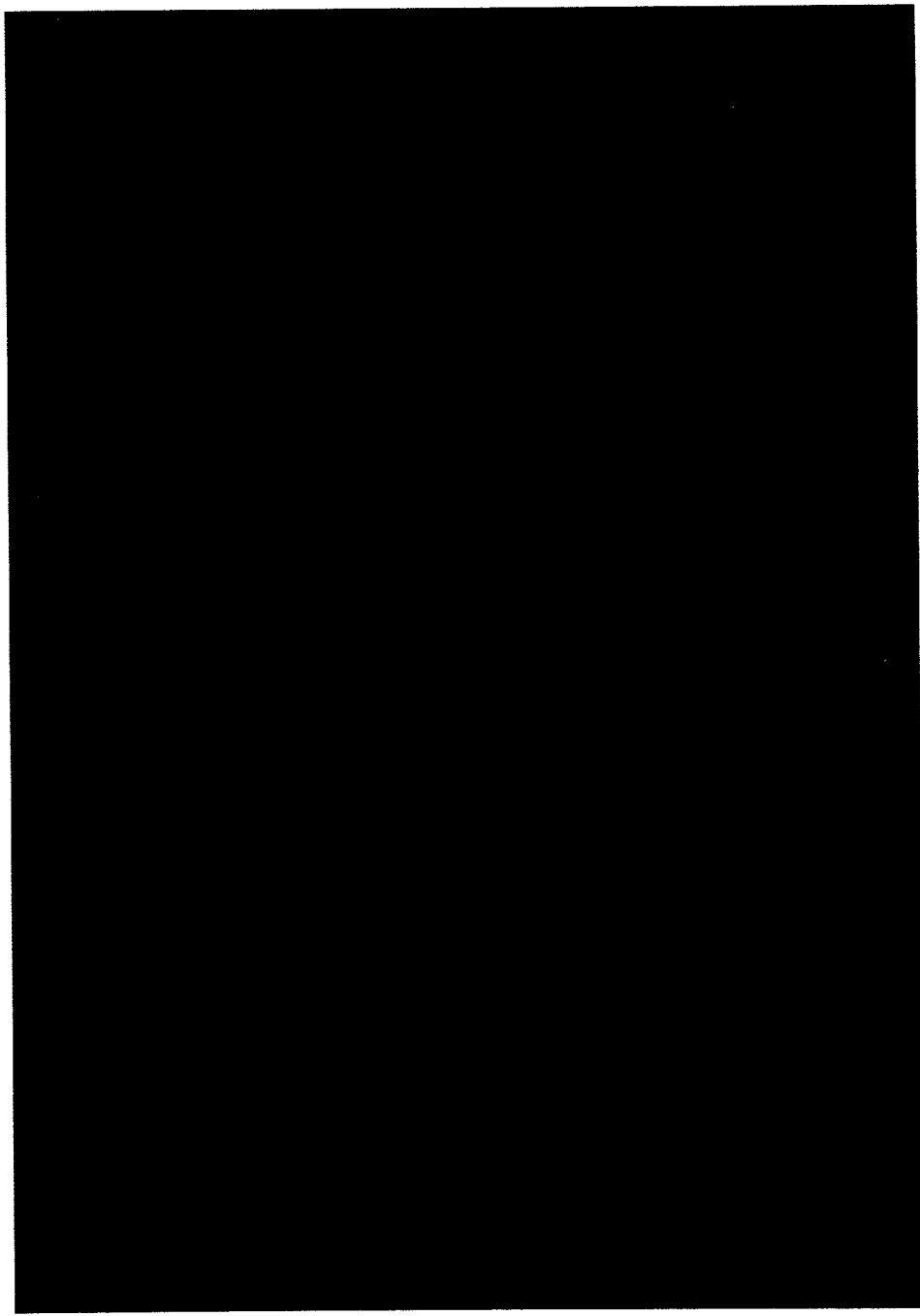
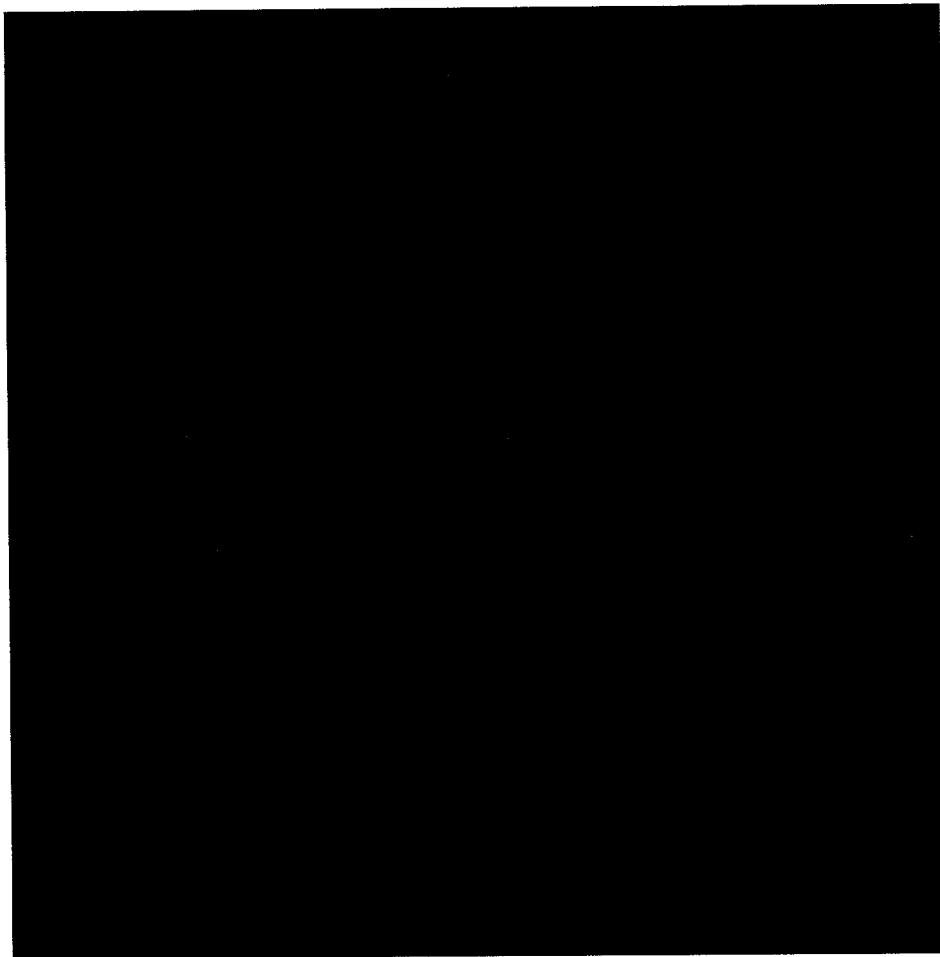
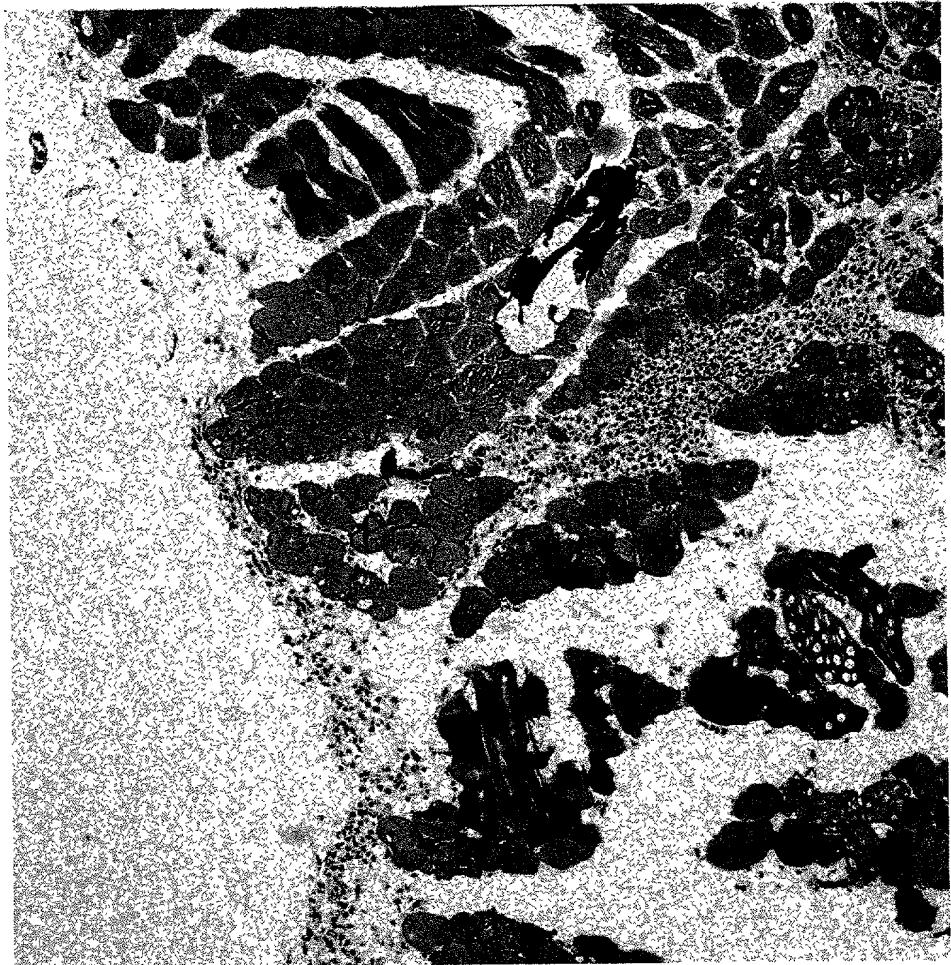


Figure 2: D3: IM injection of Dil-labeled PBMCs

D3: IM injection of Dil-labeled PBMCs



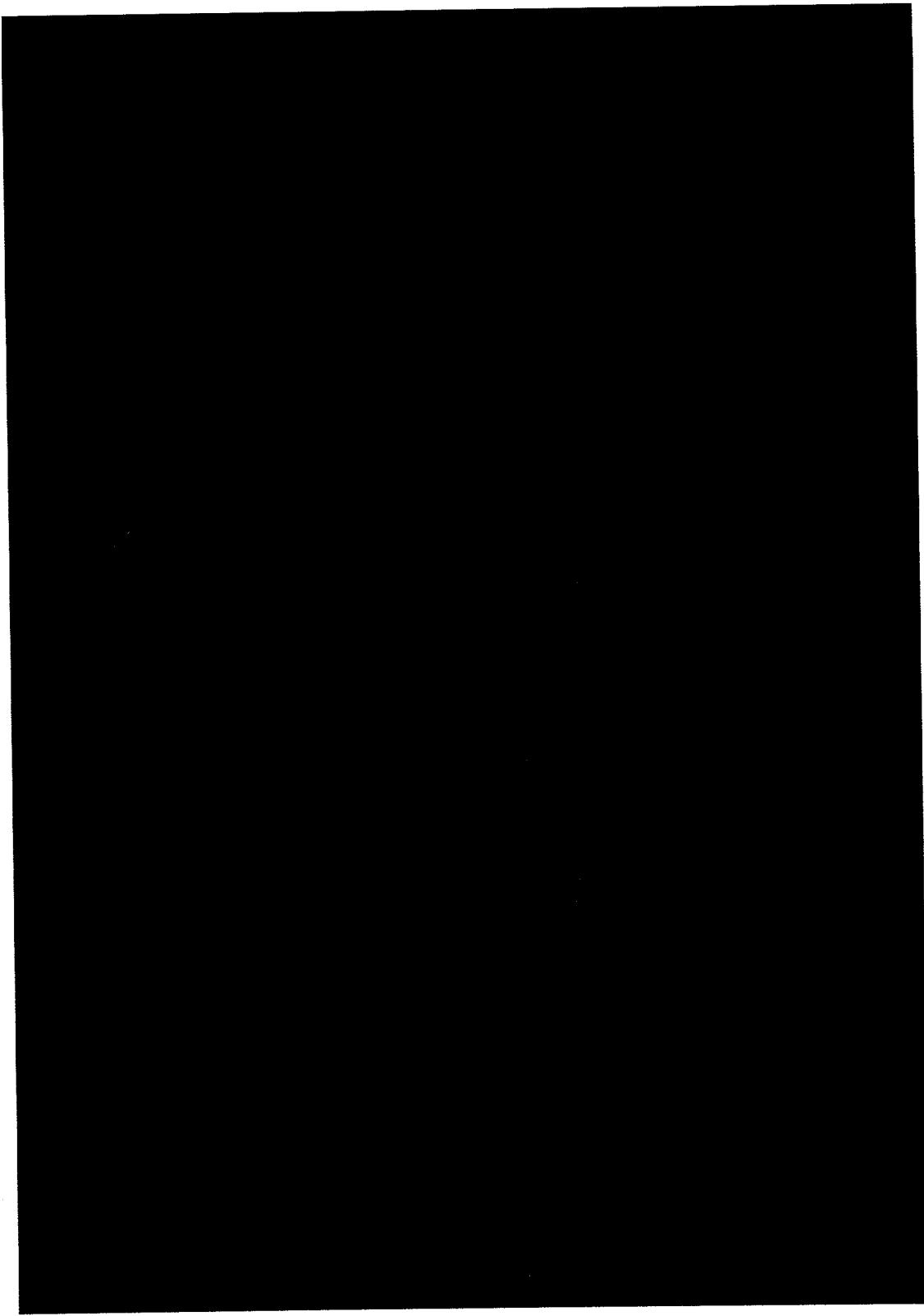
卷之三

Figure-3

D3 IM injection of saline



Fig. 4 6-Week DAPI labeled hMSCs in Mouse Muscle



Red and Blue Fluorescence Channels merged

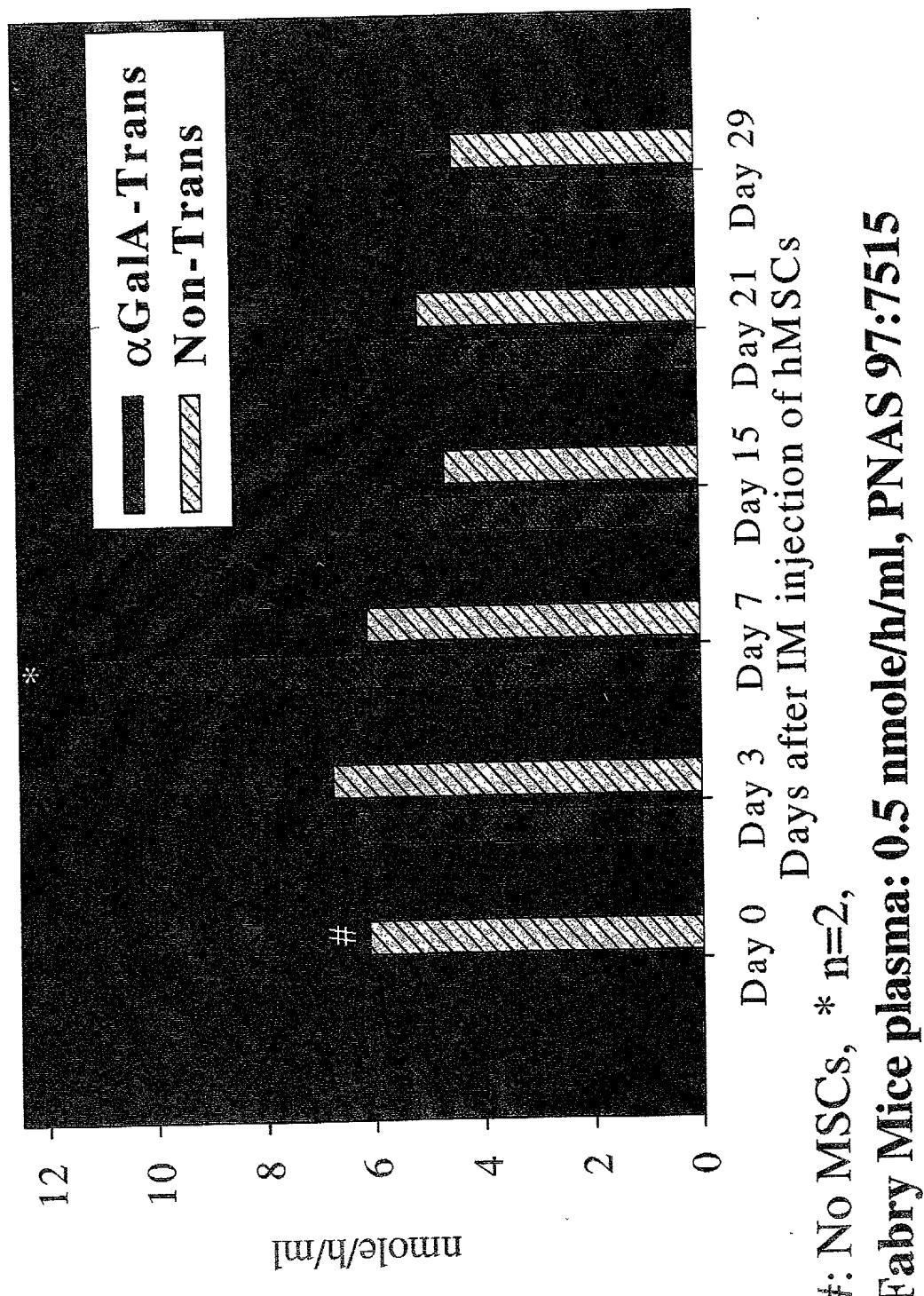
Fig.5

α GalA Immunostain of Muscle from Mouse
injected IM with α GalA-hMSCs: 14 days



Figure 6

α -GalA activity in Plasma of Mice IM-injected with α -GalA-Transduced hMSCs



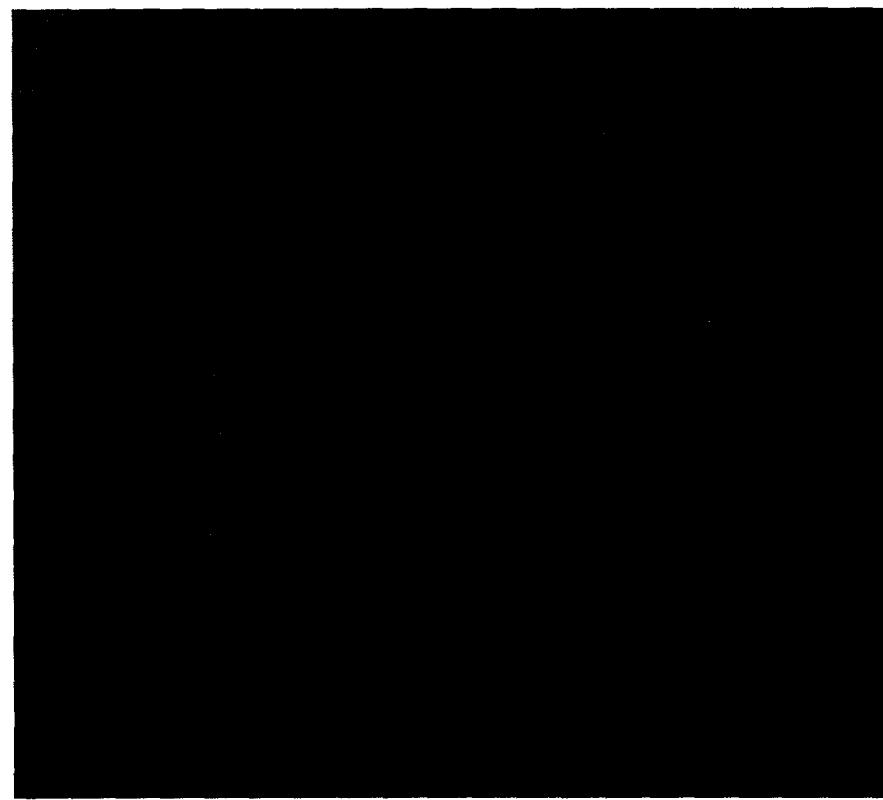
#: No MSCs, * n=2,

Fabry Mice plasma: 0.5 nmole/h/ml, PNAS 97:7515

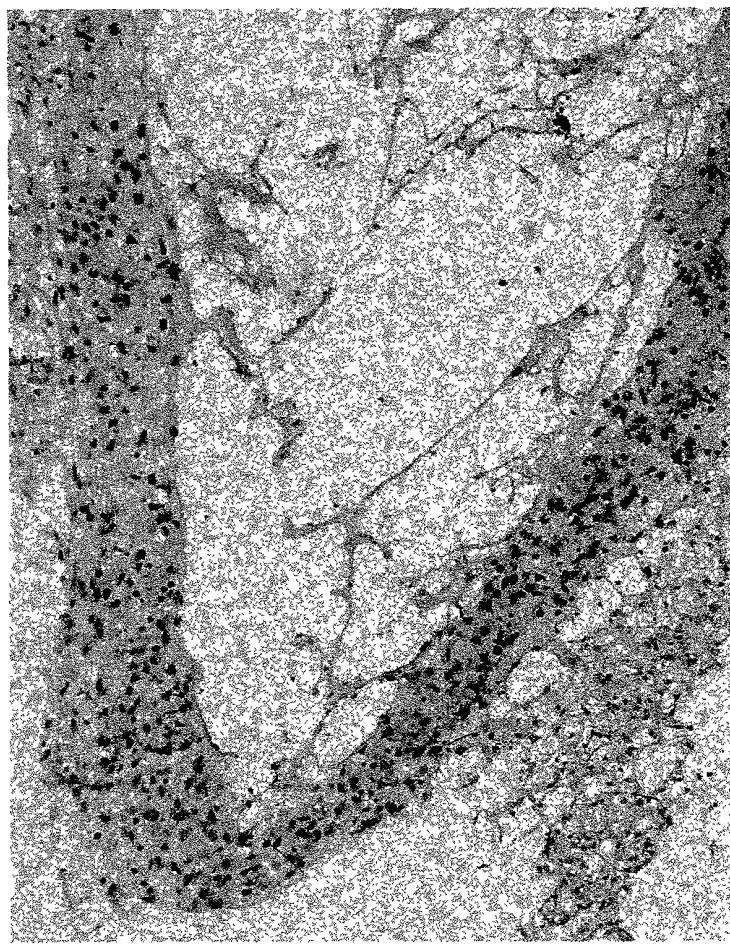
Figure 7a

Human-specific Alu sequence staining of hMSCs injected into Immuno-competent mouse muscle

Dil-positive cells

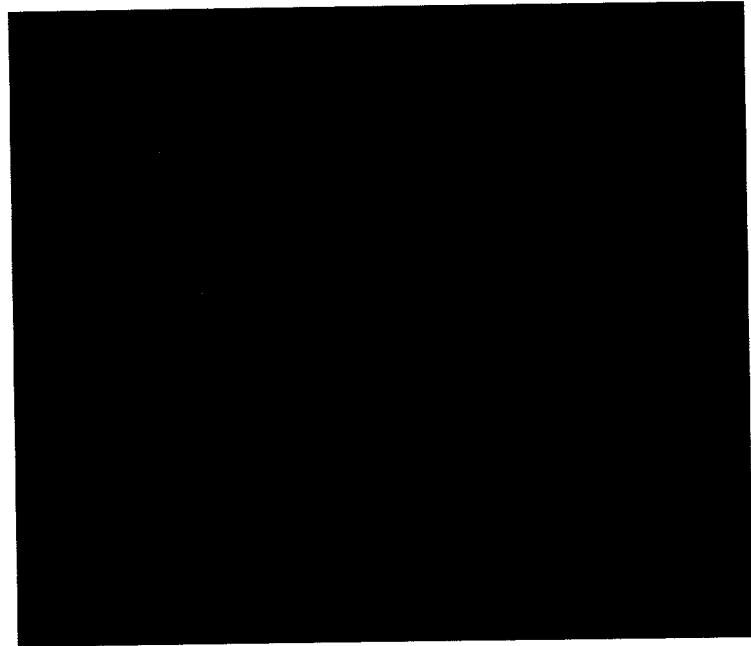


Alu-sequence positive cells

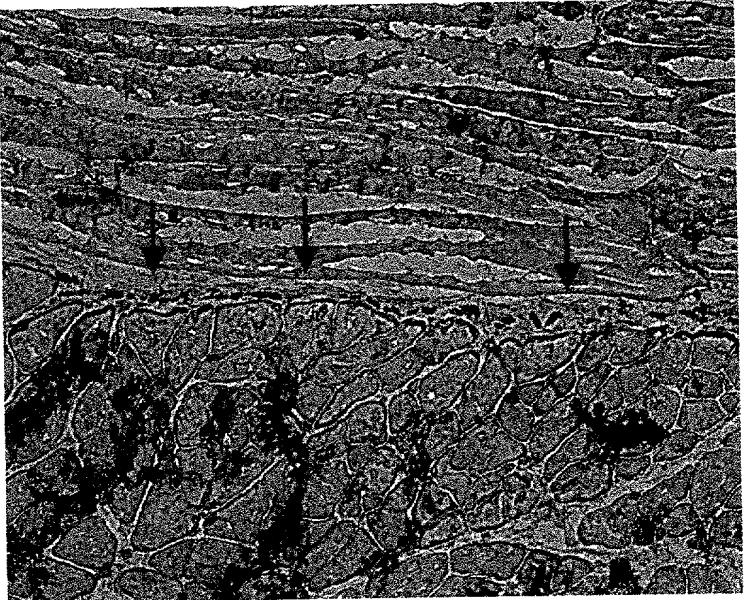


Human MSCs in Mouse Muscle: 43 days

Dil Positive Cells



Alu-Positive cells



H & E

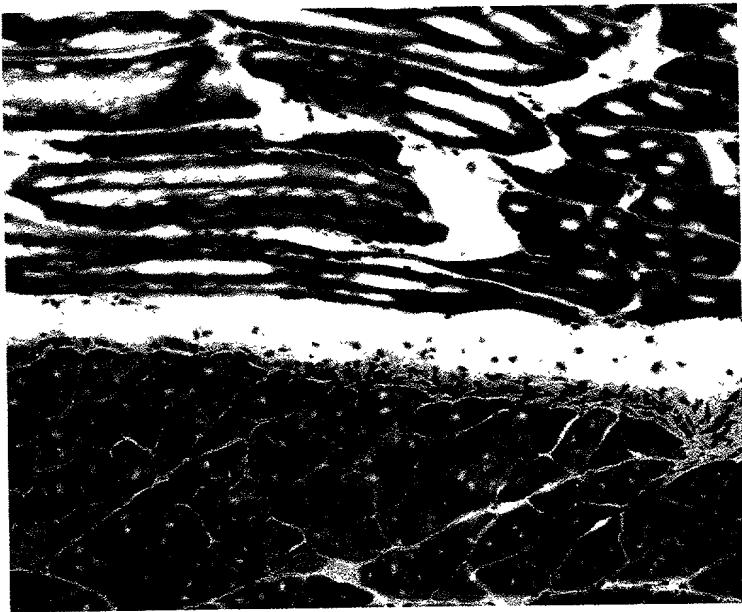


Figure 7c Day 57 DiI/Alu

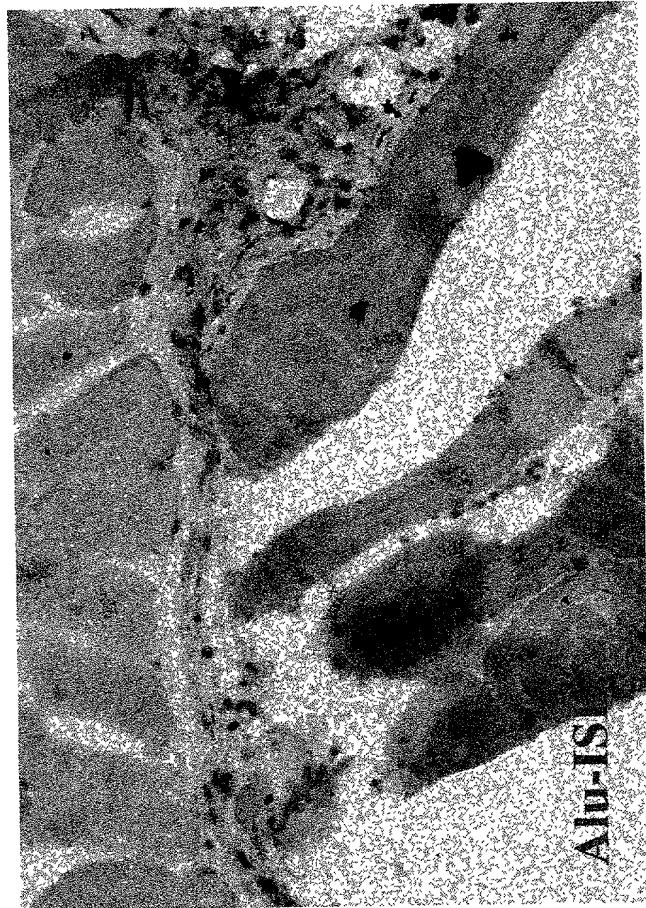
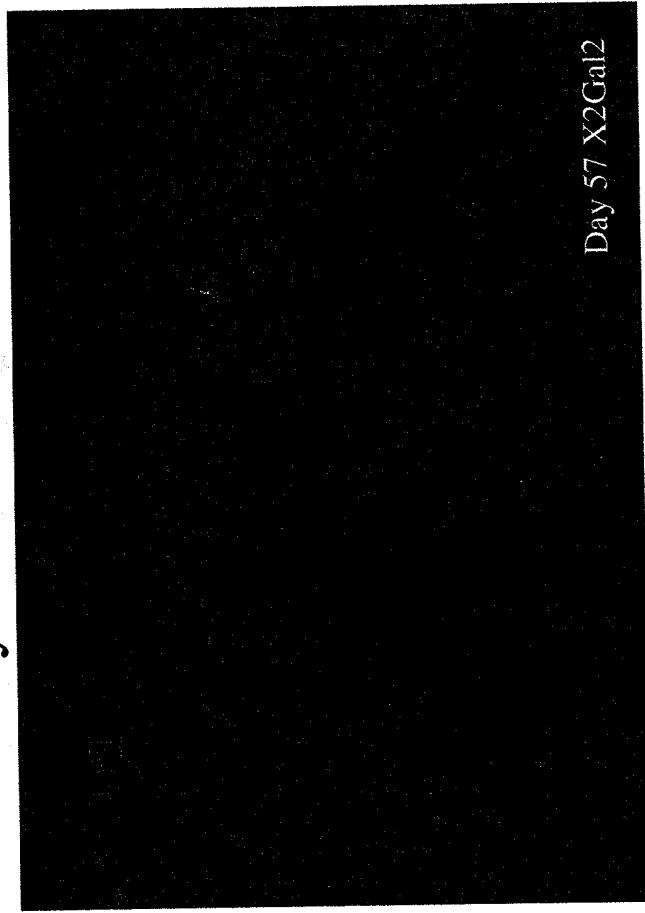
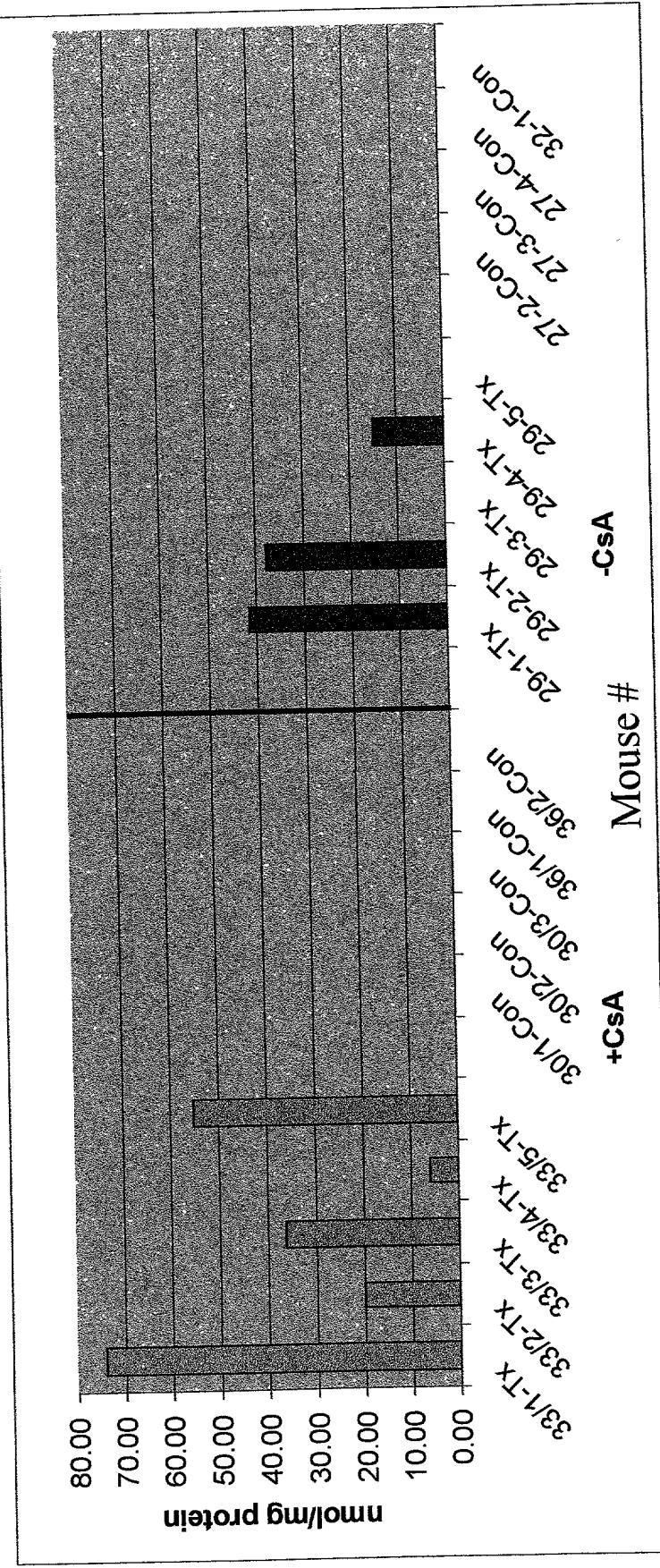


Figure-8a

α GalA Activity in Muscles of Fabry-KO mice:
14 days after iM injection of α GalA-Transduced hMSCs



$T_X = \alpha$ GalA Transduced cells injected into muscle

Con = Vehicle alone injected into muscle

Figure 8b α -GalA Activity in Muscles of Fabry-KO mice^a
28 days after iM injection of α -GalA-Transduced hMSCs

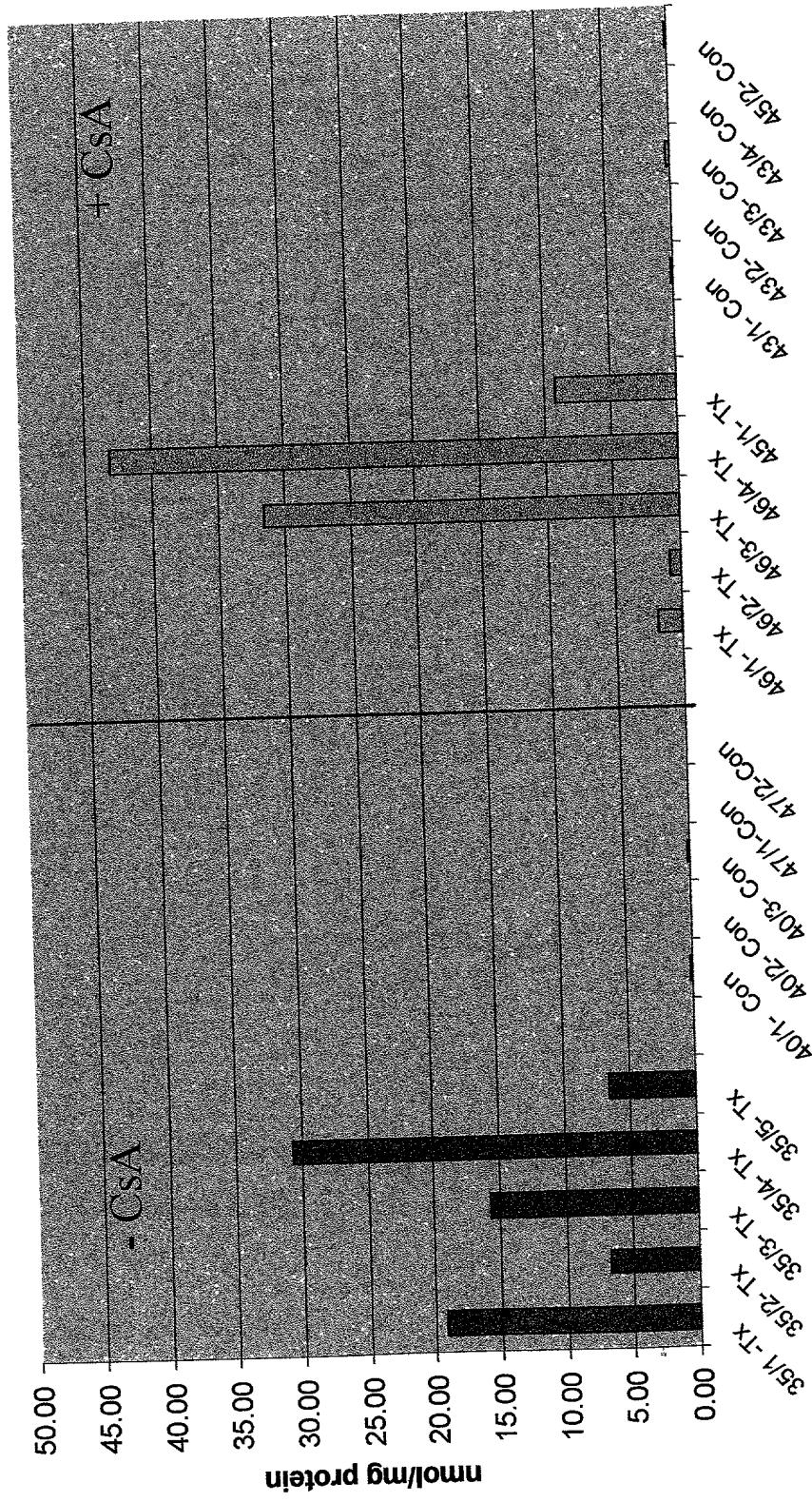


Figure-**fa**

α GalA Immunostain Of Liver From KO Mouse
injected IP with α GalA-hMSCs on Cultispheres :
14 days

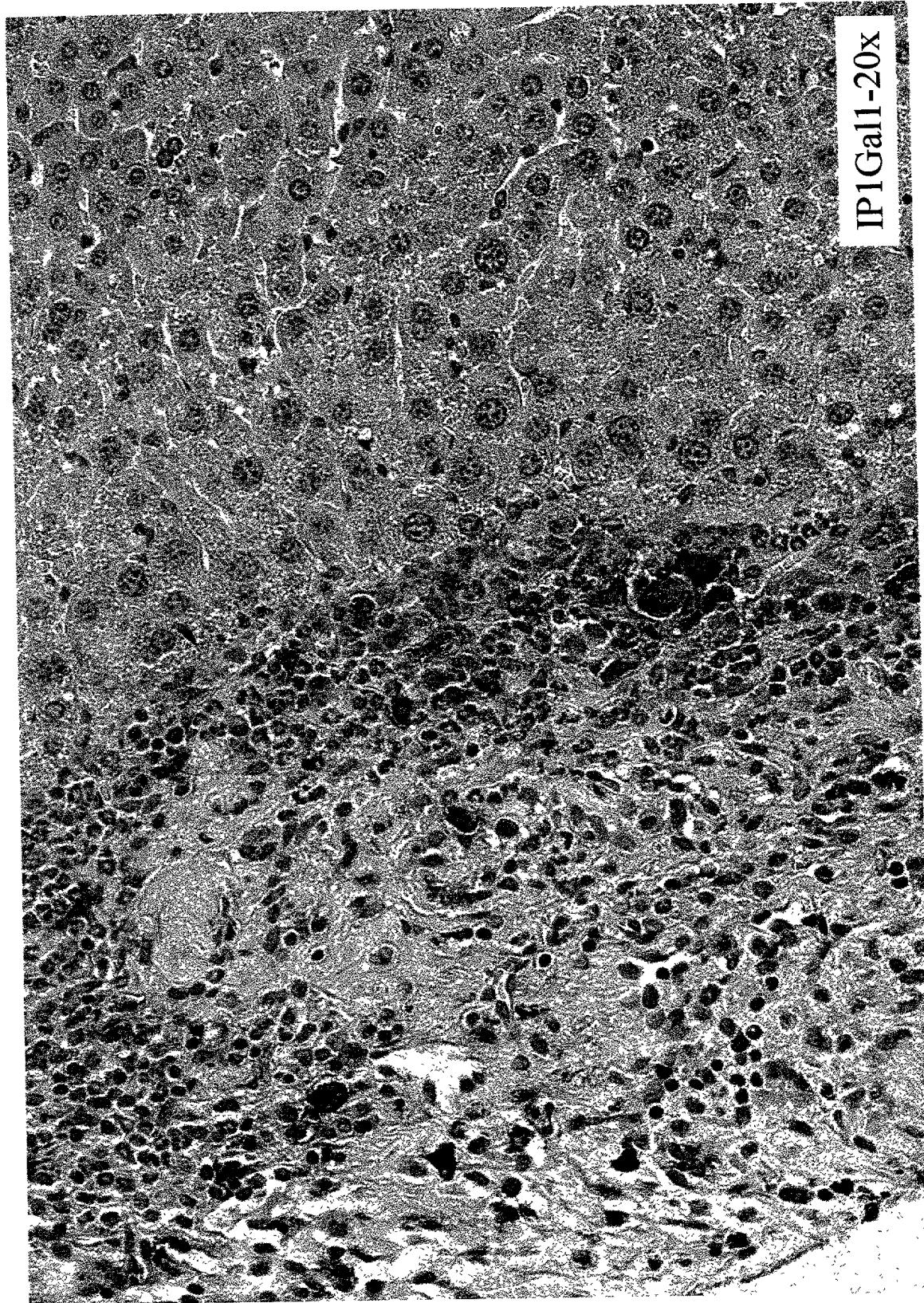


Figure 9b

α GalA immunostain of E7477f/fm KO Mouse
injected IP with α GalA-hMSCs on Cultispheres
14 days

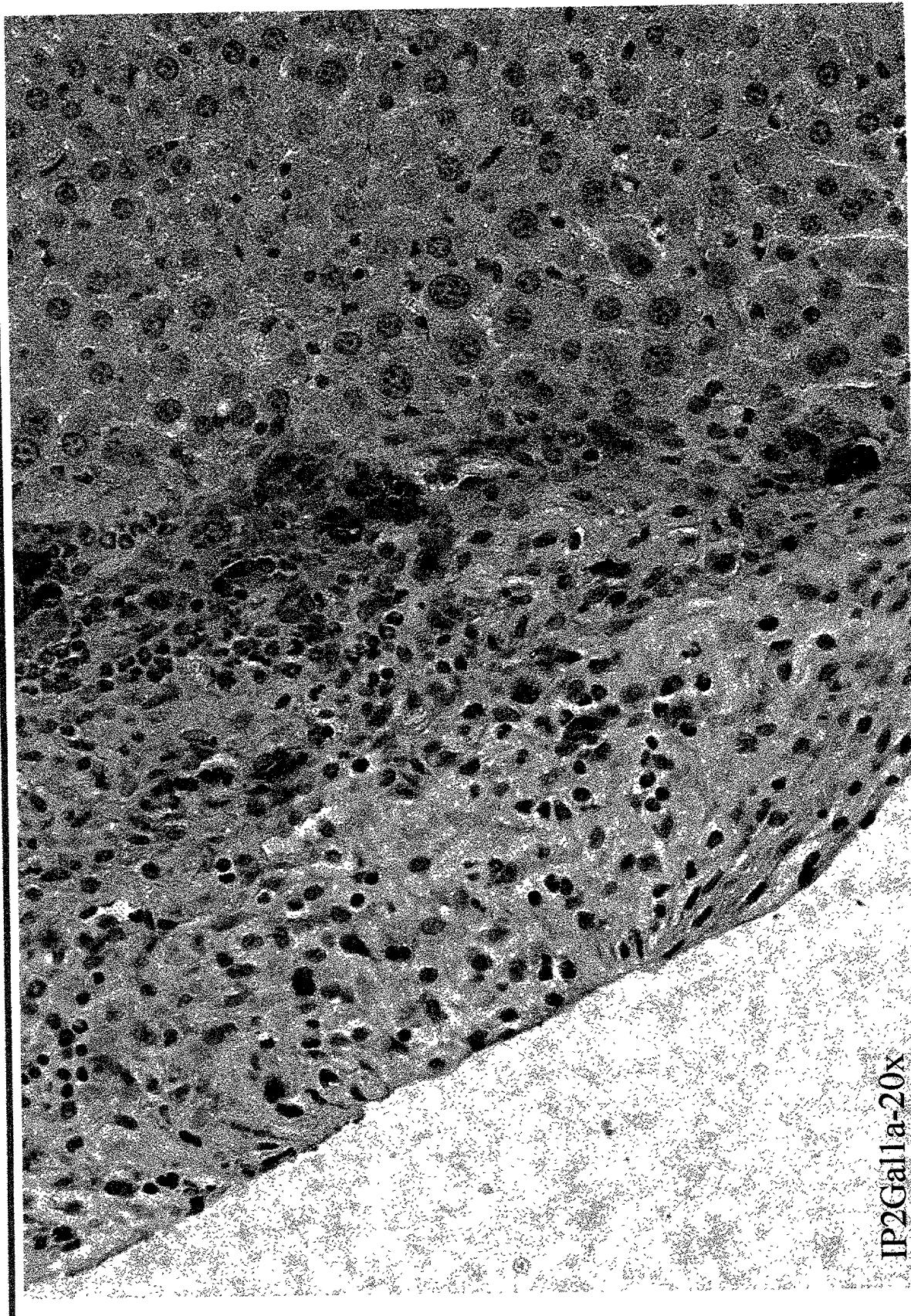
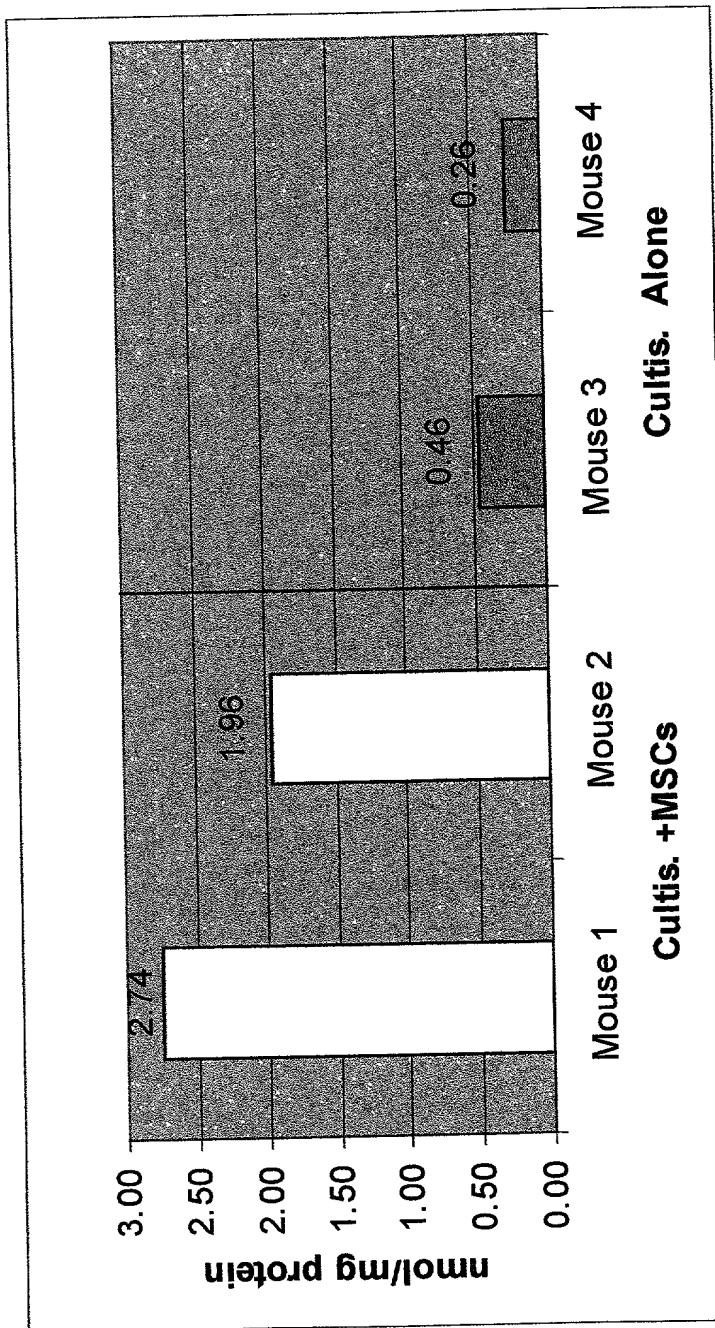


Figure 10

Intra-Peritoneal Delivery of α GalA-Transduced hMSCs to Fabry Knock-Out Mice

α GalA Enzyme



Increase in α GalA is 5% of normal mouse liver activity (41 nmole/mg)